International Journal of Contemporary Pediatrics
Ali M et al. Int J Contemp Pediatr. 2025 Jul;12(7):1208-1211
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20251767

Case Report

A case of neonatal lupus presenting with myocardial dysfunction with complete heart block: early identification and management results in excellent outcome

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Received: 16 May 2025
Revised: 01 June 2025
Accepted: 02 June 2025

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ABSTRACT

Neonatal lupus erythematosus (NLE) is an uncommon acquired autoimmune disorder that develops when maternal autoantibodies to Sjogren's syndrome A or B (SSA-SSB) autoantigens pass through placenta and cause immune mediated destruction of targeted fetal and neonatal tissues. Among the cardiac abnormalities, congenital complete atrioventricular block (AVB) is the most common cardiovascular abnormality seen in affected fetuses and infants that also accounts for more than 80% of cases of complete AVB in newborns with structurally normal heart. Other cardiovascular manifestations may include arrhythmias, myocarditis, cardiomyopathy and structural heart defects with particular tendency toward valvular lesions. We report a case of a neonate who presented with severe ventricular dysfunction with ejection fraction 20% in the presence of complete heart block (CHB), required extracorporeal membrane oxygenation (ECMO support). After treatment with pulse steroids and IVIG, there was a dramatic improvement in ejection fraction 55%. ECMO support was successfully weaned, and a permanent pacemaker was subsequently inserted. The neonate was discharged home in good health.

Keyword: Neonatal lupus, Congenital complete atrioventricular block, Ventricular dysfunction, Myocarditis

INTRODUCTION

Neonatal lupus is a rare, acquired autoimmune disorder resulting from the transplacental passage of maternal autoantibodies, particularly anti-Ro/SSA and/or anti-La/SSB, which can affect multiple fetal organs. The condition is primarily characterized by transient dermatological findings and the potential development of congenital heart block (CHB).[1] Congenital heart block (CHB) represents the most serious and distinctive cardiac manifestation of neonatal lupus, with an estimated prevalence of 1%–2% among pregnancies in women who test positive for anti-Ro antibodies.[2] Mothers with anti-Ro/SSA and anti-La/SSB antibodies have an approximately 2% risk of giving birth to a neonate with myocardial calcification, cardiac dysfunction, myocarditis, or conduction abnormalities such as heart block.[3,4] Fetal echocardiography has allowed the prenatal diagnosis of CHB, any structural cardiac defect, or myocardial dysfunction to be made routinely.[5] We present a rare case of a neonate who presented with severe ventricular dysfunction with ejection fraction 20% in the presence of CHB, required ECMO support. After treatment with pulse steroids and IVIG, there was a

dramatic improvement in ejection fraction. A written informed consent was obtained from the parents of patient for publication of available data.

CASE REPORT

A 36-year-old mother, gravida 4 para 3, known case of systemic lupus erythematous and was on Plaquenil as treatment for SLE. She received Plaquenil, aspirin, and prednisone during pregnancy. She and her husband are consanguineous. Previous baby has also congenital heart block for which permanent pacemaker was inserted. Antenatal echo showed bradycardia with CHB, right atrium and right ventricle dilation, impaired ventricular contractility and trivial pericardial effusion with no underlying structural heart defects or malformations. she was on regular antenatal follow up. The baby girl was delivered electively by caesarean section at 37 2/7 weeks. Her birth weight was 2.8 kg and APGAR scores were 8,8 and 9 at 1 min, 5 min and 10 min, respectively. At birth her tone and reflexes were good and heart rate was reported 80 bpm. After shifting to Neonatal intensive care unit (NICU), she started to have desaturation which didn't respond to CPAP so she was intubated due to persistent hypoxia. The chest radiograph showed moderate cardiomegaly and pulmonary edema. A 12-lead ECG demonstrated complete AV block. Bedside echocardiogram ruled out any structural heart defects, and showed moderate to severe depressed left ventricular function with ejection fraction of 40%, severely dilated Right atrium and right ventricle, severely depressed right side systolic functions, severe pulmonary hypertension, there was no echocardiographic evidence of endomyocardial fibroelastosis (EFE). Sepsis work-up was done and empiric antibiotics were given as per in unit protocol. Based upon echocardiogram findings of right side of heart and persistent hypoxia inhaled nitric oxide was started later oral pulmonary vasodilator also started. Given the maternal history of SLE, antenatal findings of CHB and echocardiographic findings anti-Ro (SSA) and anti-La (SSB) antibodies were sent and it came positive for it.

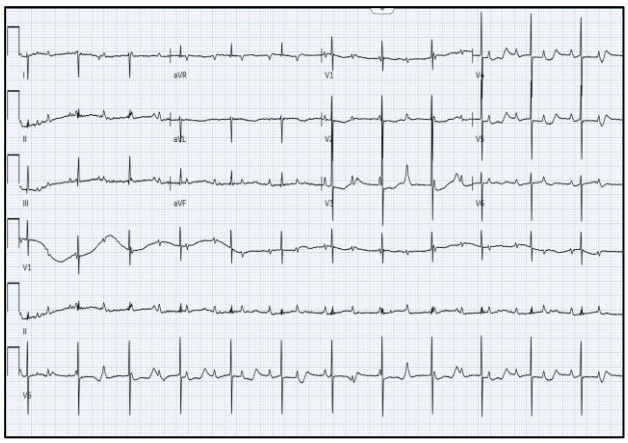

Figure 1: ECG: complete AV block.

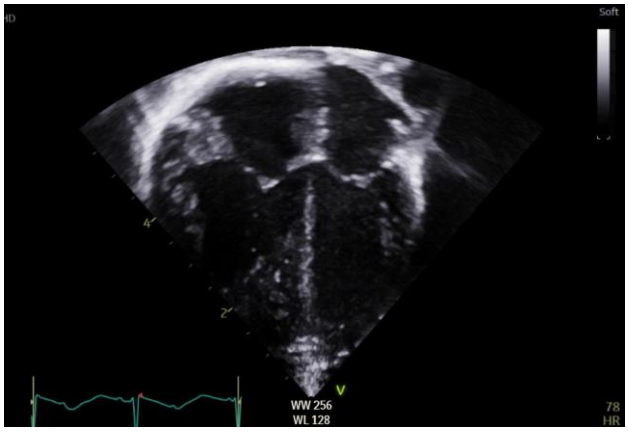

Figure 2: Four chamber echocardiographic view showing normal heart structure, dilated right atrium and right ventricles.

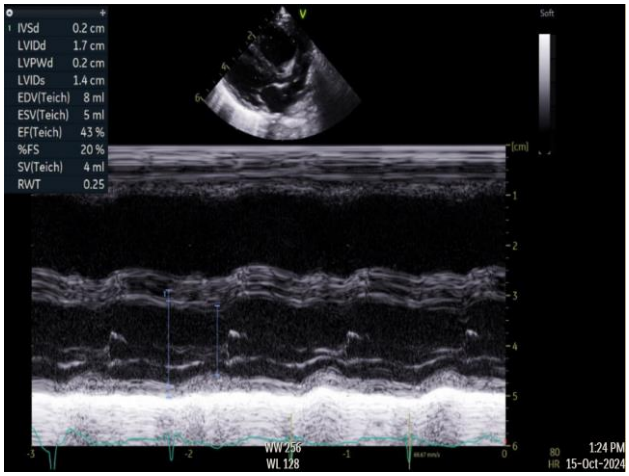

Figure 3: Parasternal long axis view showing mild to moderate depressed LV function.

On day of life (DOL) 2, she started to deteriorated with clinical findings poor perfusion, hypotension, increasing requirement of inotropes, increasing trend of lactate that reached 9, myocarditis was suspected and pediatric rheumatology was involved. she was shifted to cardiac surgical intensive care unit (CSICU) for further management and insertion of temporary pacemaker. Echo was repeated again and ejection fraction further reduced to 14% only with severe biventricular dysfunction suggesting myocarditis. Considering Immune mediated myocarditis that led to myocardial dysfunction and low cardiac output after involving Pediatric rheumatology steroids and IVIG started. Patient went OR in critical condition for pacemaker insertion and temporary pacemaker was inserted with rate of 130 bpm VVI. Even after pacing with temporary pacemaker her clinical condition didn't improve and signs of low cardiac out persists. ECMO was initiated considering very high inotrope requirement and high lactate. after placing on VA central ECMO lactate improved and her clinical condition started to get better. She remained on ECMO for 4 days to rest the myocardium to recover and for

treatment of myocarditis inform of IVIG and steroids to work. On day 06 (DOL) echo was repeated and it showed significant improvement in EF 55%. 0n day 07 (DOL) ECMO support was weaned and she was decannulated successfully. On day 08 (DOL) permanent pacemaker was inserted with rate of 120 bpm.

On day 15 (DOL) he was extubated on CPAP weaned to nasal cannula and later finally room air. On day 30 (DOL) she was discharged home on room air, with oral sildenafil and bosentan. She was tapered off steroids at age of 3months with normal ACTH stimulation test.

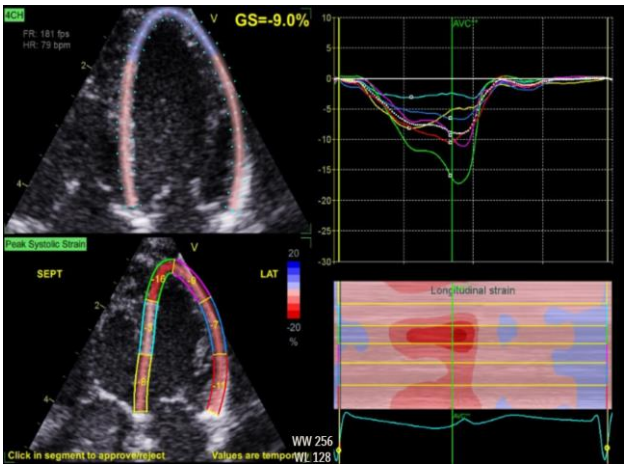

Figure 4: Tow chambers echocardiographic view showing GLS for left ventricles which is severely depressed LV function.

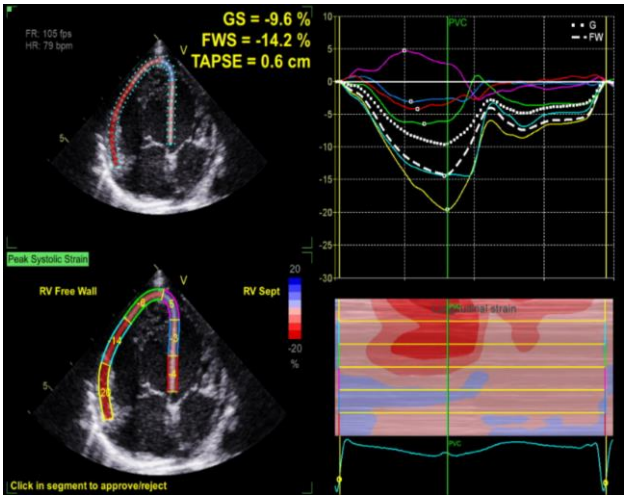

Figure 5: Four chambers echocardiographic view showing TAPSE for right ventricles which is severely depressed RV function.

DISCUSSION

Neonatal lupus is the leading cause of congenital heart block in fetus or neonates without structural heart defects.[6-10] In the majority of cases, CHB is marked by pathological changes involving fibrous tissue, which either replaces the AV node and its nearby structures or disrupts the connection between the atrial myocardium and the AV node.[11] For a long time, CHB was believed to be the only significant cardiac abnormality in newborns with high levels of anti-Ro (SSA) and anti-La (SSB) antibodies. However, accumulating clinical experience has revealed a broader range of cardiac complications associated with neonatal lupus. These include not only conduction abnormalities, but also ventricular dilation, impaired systolic function, myocardial hypertrophy, and, most commonly EFE.[12,13]

Between 15% and 20% of neonatal lupus cases show signs of widespread myocardial disease before birth, while others may develop myocardial dysfunction after birth, even when pacemaker treatment is properly administered.[14-16] This widespread myocardial disease can occur even without conduction abnormalities, indicating that cardiomyopathy might represent an independent manifestation of neonatal lupus.[14] We present a rare and noteworthy case of NLE, characterized by severe myocardial dysfunction occurring alongside CHB. Notably, the myocardial dysfunction persisted despite pacemaker insertion. However, in contrast to previous reports, our patient demonstrated a rapid and remarkable improvement in cardiac function following the initiation of NLE-specific treatment. Within just four days, the ejection fraction improved from 20% to 55%, allowing successful weaning from ECMO support. We believe that our patient's clinical condition was due to a combination of CHB and severe myocardial dysfunction, as her symptoms did not improve despite pacemaker insertion. Early NLE treatment results in complete recovery of myocardial function. To the best of our knowledge, this is the first reported case of NLE presenting with severe myocardial dysfunction alongside CHB, requiring ECMO support and demonstrating a dramatic response to pulse steroid and IVIG therapy resulted in full myocardial recovery. This case expands the known clinical spectrum of NLE and demonstrates the importance of early recognition of NLE-induced myocarditis. Prompt initiation of appropriate treatment led to an excellent clinical outcome.

CONCLUSION

NLE can present with both congenital CHB and myocardial dysfunction inform of myocarditis**.** Prompt recognition of suspected neonatal myocarditis and early initiation of pulse corticosteroids along with IVIG therapy can lead to markedly favorable outcomes.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Ali M, Beheri RM, Alheraish YA, Almuqri R, Alarwan KMN, Alomair MAM, et al. A case of neonatal lupus presenting with myocardial dysfunction with complete heart block: early identification and management results in excellent outcome. Int J Contemp Pediatr 2025;12:1208-11.